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ON THE CLASSIFICATION OF THE CLINICAL FORMS OF TULAREMIA

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Both clinicians and epidemologists have long maintained the need for a classification of the clinical forms if tularemia. Such a classification must reflect the clinical-pathogenetic and epidemological aspects of this disease, and also promote the establishment of dependence between the clinical manifistations and epidemology of each individual case and each outbreak of tularemia.

The classification proposed by American authors was arranged purely empirically, and the forms included in it glandular or bubonic, skin-glandular or ulcer-bubonic, eye-glandular and typhoid) did not reflect the full diversity of clinical manifiestations of tularemia. A number of Soviet authors (Khatenever, Gayskiy, Bilibin, Berinskaya, Rudnev, Gromaskevskiy, Mayskiy, and others) who have made vlauable contributions to the study of the pathogensis, clinical manifistations, therapy, and specific prophylaxis of tularemia, also proposing their own classifications, but not one of these received general recognition. At the All-Russian conference on tularemia the division of the disease into two basic forms was accepted: glandular (bubonic) with symptoms of local bubos, and septic ("generalized", according to Rudnev), corresponding to the previous so-called "typhoid" form.

However, subsequent deepening and widening of our knowledge about tularemia has shown the improcticality of differentiating septic form. Studies of the pathogenesis and pathological anatomy

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of this disease have completely established that there is no tularemia without a subsequent consummation of the pathological process with a bacteriemic phase. To isolate this phase is just as irrational, as, for instance, to create a special form of typhus based on the phase of bacteriemia which is inevitably present with it. Against this we have not only the unfailing discovery of pathological changes in lymph node sections of those cases in which a clinical diagnosis of the septic form was made, but also the absence, in patients, of the syndrome characterizing the septic reaction which is well-known to every clinician. It is known how frequently patients with a diagnosis of the septic form of tularemia feel so well that they do not even require hospitalization.

Experience made it necessary to reconsider the question, and in 1946 the All-Russian conference on tularemia gave a new ckubuca form of classification of this disease: (1) bubomic (glandular) with indication of the localization, (2) visceral (internal), subdivided into: (a) bronchiO-pulmonary (lung) and (b) abdominal (intestinal).

At that stage of our knowledge concerning tularemia the term "visceral" was progressive, since it disclosed the nature of the disease as a general process, sometimes manifesting itself as damage to internal organs, refuting, in this way, the existence of an independent "generalized" form, and facilitating the formulation of a more modern view of the pathogenesis and clinical symptoms of tularemia. Subsequently this term too ceased to satisfy our practical needs, since the presence of the visceral form sometimes allowed the doctor to avoid a closer internal localization, while such

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localization is very important from the epidemological point of view. Furthermore, the classification accepted accepted by the All-Russian conference on tularemia was not obligatory for the other republics of the Soviet Union, which led to an extraordinary diversity of reports presented to the Ministry of Public Health USSR, making it almost impossible to draw conclusions from the information in them. In view of this, in 1950, after a series of preliminary conferences with specialists in tularemia, the ministry of Health USSR, proceeding from the position that tularemia is a general disease, in which the development of all pathophysiological processes is regulated by the central nervous system, and that the clinical aspects and anatomy of this disease appear with primary lesions in the region of the entrance gateways of the infection, with a necessary presence of regional lymphadenitis, affirmed the following classification of the clinical forms of tularemia.

According to the localization of the process: (A) tularemia with external symptoms of pathology in the skin, mucous membranes and lymph nodes: (1) bubonic, (2) ulcerative-bubonic, (3) ocular (ocular-bubonic), (4) anginous-bubonic, (5) with the presence of other external localizations; (B) with pathology mainly of the internal organs: (1) respiratory system, (2) gastro-intestinal tract, (3) with affliction of other internal organs.

According to the severity of the process: (1) light, (2) severe, (3) average severity.

According to duration: (1) acute, (2) prolonged, (3) recurrent.

In this classification there are the three following principal new points.

1. Exclusion of the septic and primary pulmonary forms, whose existence is not supported by sufficiently convincing pathological-anatomical, clinical and epidemological evidence.

2. A dynamic classification: the presence of categories, which allow for internal or external localizations of the process, which show that the study of tularemia is not yet completed, and give ample scope for the

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initiative of future investigatiors, not obliging them to force newly described localizations into the framework of a fixed scheme.

3. Finally, the given classification allows each case to be examined in three planes: the localization of the process and, consequently, the way of entry of infection; the severity of the process; and its dynamics. The first of these points is particularly important, since tularemia belongs to those diseases in which the correct explanation of the localization of the process always accurately indicates the entry path of the infection; an epidemologist can determine the origin of the latter almost without mistake.

In order to make the use of the classification easier for the practicing doctor, supplementary instructions were attached to it to clarify by what symtoms one or anothe case can be included in this or that clinical form. The basic goal of the instructions is to help the doctor in a correct clinical diagnosis of tularemia and to establish a link between the clinical manifistations and the epidemology of each individual case of this disease.

According to these instructions, the bubonic form includes cases in which the infection enters through the skin, leaving traces in it during the process in the form of a primary affect, and leading to the development of regional lymphadentites. This form of tularemia is present in commercial outbursts.

In opposition to this form of transmission, superficial ulcers characteristically arise at the point of penetration of the infection, with a subsequent spread of the infectious process (bubos) to the nearest lymph nodes (ulcerative-bubonic form).

The ocular or ocular-bubonic form in which the infection penetrates through the mucous membrane of the eyelids, usually in threshing epidemics, but sometimes also in outbreaks of everyday Origin.

The anginous-bubonic form appears most regularly as a result of alimentary mode of infection. It expresses itself in specific angina and inflammatory reactions of regional lymph nodes.

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The alimentary form of infection also brings about that form which is called tularemia of the gastro-intestinal tract in the classification. The primary pathologo-anatomical substrate in this form of tularemia, as in any form, is the bubo, which may develop in any lymph node along the path of the gastro-intestinal tract where infection penetrates. These buboes are sometimes very small and not capable of palpation, and therefore this form is distinguished by a sparsity of local clinical symptoms.

The same is also characteristic of the second internal form, manifested in pathology of the respiratioy tract. It appears as a result of infection by the aspirative path, and it is therefore natural that it is most common in outbursts of a threshing origin. The difficulty in diagnosing the two latter forms was the resaon for the diagnosis of them, in particular, first as typhoid and then septic forms. By depending on a correct epidemological analysis of every case, the clinician will make it much easier for himself to deagnose tularemia that manifests itself as an affliction internal organs.

In considering the question of the severity of the tularemia, one must always proceed from the position, which has already been presented above, that tularemia, regardless of its localization, is a general disease, in which all development of pathological processes in the organism is regulated by the central nervous system. Ror this reason, the severity of the disease is determined first of all by the degree of general intoxication. In correspondence with this formulation, the severe form should include cases accompanied with a sharply defined intoxication, depression of the central system, disturbance of the functional condition of the heart and vessels, and associated complecations, which present a threat to the life of the patient. On the other hand, one should consider as light cases those in which there is no marked intoxication and the patient maintains satisfoctory health during the course of the whole pyretic period, which continues for no more than seven days. Cases of average severity occupy an intermediate position between the light and sever.

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A primary criterion for determining the duration of tularemia, in the instructions, is the time starting with the appearance of clinical symptoms of the disease. Thus, the acute form should mean that form of tularemia which started suddenly and continued for two to three months. Chronic or prolonged tularemia is characterized by a maintenance by the disease of all the typical symptoms for a period of up to a year, and sometimes even longer. Recurrent tularemia is taken to mean a form in which a man, who recovered one to two years previously and is practically well, once again shows the clinical symptoms of this disease syndrome, under conditions where the possibility of reinfection is excluded.

Let us hope that the instructions will orient practicing doctors correctly, helping them to understand the accepted classification, and aid in the analysis of the clinical aspects and epidemology of each individual case and each outburst of tularemia. On the other hand, this unified approach to the data on tularemia outbursts makes them suitable for epidemological analysis and will make possible a systematization and use of accumulated experience in the battle with tularemia.

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